

274* RhDNase before or after going to sleep in children with cystic fibrosis?

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Objective: To compare in patients with CF who are on maintenance therapy with rhDNase the efficacy and possible side effects of nebulisation of rhDNase before sleep to nebulisation in the morning.

Methods: Design: randomized, double blind, double dummy, cross over study. Inclusion criteria: CF, stable clinical condition, rhDNase maintenance therapy. Children in Group I inhaled rhDNase before sleep, and a placebo in the morning in week 1–2. The protocol was reversed during week 3–4. Group II performed the reversed sequence. Patients continued their daily routine sputum expectoration. Primary endpoint: MEF25. Pulmonary functions tests were performed on day 0, 7, 14, 21 and 28. After 1, 2, 3 and 4 weeks oxygen saturation and cough frequency were measured during the night.

Results: 24 patients completed the study. Mean age: 13 years (range 6–19). MEF25 (%pred) as primary endpoint did not show a significant difference between nebulisation of rhDNase before sleep versus nebulisation of rhDNase in the morning. Nocturnal cough, oxygen saturation, or any of the other secondary end points were not significantly different between the two study periods.

Conclusion: This study showed in children with CF lung disease who are on maintenance treatment with rhDNase that it is equally effective and safe to nebulise rhDNase before going to sleep compared to nebulisation in the morning.

276 Perception versus objective assessment of diseases severity in cohort adult cystic fibrosis (CF) patients

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Introduction: The aims of this research is to investigate whether the degree of disease severity as assessed by adult CF patients (perception) correlate with recognised CF outcome measures. This was compared with an objective performance score.

Patients and methods: We investigated 76 adult CF patients (29 female), mean age 26.6, range 17–42 years. At their 2006 annual assessment, perception of health score (PHS) was estimated by patients' scoring their own health status. This was graded on a scale 0–10. Objective performance score (OPS) was estimated by summation of 4 domains (score 1–5 in each). These were: breathlessness score, chest examination score, chest radiograph score and BMI score. This system was validated: (Middleditch AG, Jarad NA. *Am J Respir Crit Care Med* 2003;167:A499). Factors assessed for comparison were: age, sex, FEV1, annual number of pulmonary severe exacerbations (P Exs) and treatment burden.

Results: Out of 76 patients 64 (84%) OPS and 56 (74%) PHS was available for comparison. There was no difference between female and male patients with regards to either score and neither scoring system correlated with age. OPS correlated with PHS, $r=0.63$, $P<0.0001$. Both scores correlated with percentage predicted FEV1 although OPS correlated more closely than PHS, $r=0.79$, $P<0.0001$ for OPS and $r=0.53$, $P<0.0001$ for PHS. Both scores inversely correlated with the number of P Exs ($r=-0.33$, $P=0.013$ for PHS) and ($r=-0.54$, $P<0.001$ for OPS), and with the treatment burden.

Conclusion: We found that both, own health score and objective performance score correlated with known outcome measures in patients with cystic fibrosis. There was a closer correlation for performance score.

275 Questionnaire survey of stress urinary incontinence (UI) in 9–16 year olds with cystic fibrosis, compared to other chronic respiratory conditions and a normal group

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Aims: To report incidence, degree and impact of UI in 9–16 year olds with CF, compared to controls and clinical status.

Method: Children aged 9–16 years, from clinics at a tertiary centre. Group 1: CF clinic. Group 2: Respiratory clinic (chronic cough-asthma, bronchiectasis or PCD); main cause of UI in CF is cough [1]. Group 3: Normal control (fracture clinic). At clinic – consented before self administered questionnaire (Group 3: short anonymous version). Data collected: sex, age, % predicted FVC & FEV1, height, weight and BMI z scores. Group 3: age & sex only.

Results: Little difference in baseline data, except Group 1 were shorter ($p=0.071$). No statistically significant difference between incidence of wetting, on more than 1 occasion. Laughing, exercise & cough were the most common causes of UI. No difference between wetters for age range, lung function, physiotherapy, breathlessness, antibiotics, urinary tract infections, menarche. Only 3 (group 1) and 2 (group 2) report more than a few drops of UI.

Conclusion: No difference was shown in incidence of UI between the groups. Reported incidence is similar to that of other studies without control groups in CF children, but also in healthy adolescents [2].

References

[1] Dodd et al. *J R Soc Med*. 2005; 98(Suppl 45): 28–36.

[2] Swithinbanks et al. *Br J Urol*. 1998; 81(Suppl 3): 90.

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	Group 1: CF (N=81)		Group 2: Respiratory (N=55)		Group 3: Normal (N=58)	
	Girls	Boys	Girls	Boys	Girls	Boys (2 unknown)
Numbers	36	45	21	34	28	28
Age (Range)	12.1 (9.2–16.9)	12.6 (9.0–16.7)	12.3 (9.6–16.2)	12.9 (9.0–16.9)	11.3 (9.2–16.1)	12.88 (9.6–16.0)
Wetting on more than 1 occasion	11 (30%)	6 (13%)	4 (19%)	8 (31%)	7 (25%)	3 (11%)
	17 (21%)		12 (22%)		10 (17%)	

277 The effects of musculoskeletal physiotherapy and massage on pain and ease of breathing in adults with cystic fibrosis

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Musculoskeletal pain is prevalent in cystic fibrosis (CF). The aim of this study was to examine the effect of musculoskeletal physiotherapy techniques and soft tissue therapy, including massage on pain and ease of breathing (EOB) in adults with CF. **Methods:** 129 adults with CF (60 males, 70 with acute exacerbation), including 24 post lung transplant with a mean (\pm SD) age of 31 ± 9 years and FEV1% predicted of 51 ± 21 participated in this study. Following assessment of pain regions, each subject underwent a single individualized session including spinal joint/intercostal mobilisation, soft tissue therapy, exercises and postural advice of one hour duration. Subjects rated their pain levels and EOB on a visual analogue scale (VAS) prior to and at the session's conclusion.

Results: Pain was most commonly reported in the thoracic spine region (38% of subjects), followed by the shoulder region (31%), cervical spine region (16%), chest wall region (9%) and lumbar spine region (9%). EOB rating prior to treatment were worst in those with low BMI ($r=-0.21$, $p=0.02$) and low FEV1% predicted ($r=-0.24$, $p=0.01$). A single treatment session was associated with reduction in pain ($p<0.05$) and improvement in EOB ($p<0.05$), irrespective of clinical or transplant status. Improvement in pain was equivalent for all primary pain regions. However, greater improvement in EOB was evident in subjects with shoulder pain compared to other regions ($p<0.05$).

Conclusions: A combination of musculoskeletal physiotherapy techniques, exercise prescription and soft tissue therapy reduces musculoskeletal pain and improves EOB in adults with stable CF, during an acute exacerbation and post lung transplantation. Funded by the Simon Rhoden Foundation.